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Clinical psychology in haemoglobinopathies: Cost effective pathways to funding

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SICKLE CELL DISEASE (SCD) and thalassaemia are inherited blood disorders that vary significantly in their presentation and treatment but are known collectively as `haemoglobinopathies'. SCD is characterised by an abnormality in the synthesis of normal haemoglobin (De, 2005). In SCD, abnormally shaped haemoglobin are responsible for small vessel blockages which account for several symptoms and complications including acute episodes of pain ('crises'), organ failure and stroke. UK incidence of SCD is estimated to exceed 10,000, making it the most common genetic condition in the UK. It is most commonly diagnosed in African and Caribbean populations. Tha-lassaemia is a chronically recessive inherited blood disorder most commonly diagnosed in Eastern Mediterranean, Asian, and Middle Eastern populations. There are several different types of thalas-saemia, which vary in terms of illness severity and treatment intensity. The main types, alpha thalassaemia and beta thalas-saemia, differ depending on which haemoglobin chain is affected. Beta thalassaemia major is a severe form causing life threatening anaemia which, without medical intervention by regular blood transfusions, is fatal.

Psychosocial difficulties are common in haemoglobinopathy populations (Molock & Belgrave, 1994; Vardarki et al., 2004) and neuropsychological difficulties have been widely documented in SCD (Berkelhammer et al., 2007). The importance of clinical psychology input into services for people with SCD and thalassaemia has been recognised (Sickle Cell Society, 2008; 2010; UK Thalassaemia Society, 2008). Indeed, specialist clinical and health psychology services have been developed in other centres including London, Birmingham and Manchester. At the start of this project there was no clinical psychology provision for people with SCD and thalassaemia in the Nottingham area despite a haemoglobinopathy population of approximately 140. To secure local funding in an uncertain climate, a needs assessment was completed, where we provided evidence for a service need and produced a toolkit of materials to facilitate the future post-holder's prompt integration into the service.

Groundwork

The project was registered as a service evaluation with Nottingham University Hospital's Research and Innovation Department, and was carried out by an Assistant Psychologist (AP: JL) supervised by two Consultant Clinical Psychologists (CPs: RdN & DM). Our project involved a literature review on psychological aspects of haemoglobinopathies, information gathering from staff and service users about the current service and their needs, introducing clinical psychology to the haemoglobinopathy team, and offering short interventions with some patients. The project therefore allowed us to explore how clinical psychology was going to be conceptualised from service users and healthcare team perspectives, to evaluate the need for the service, and to examine how the service could be configured and delivered, making best use of minimal time and resources.

To introduce service users and staff to clinical psychology we attended patient involvement days and met regularly with both groups. The AP attended paediatric and adult outpatient clinics to learn about SCD, understand the key psychological issues, and explore how a CP would fit in with the service.

Provision of psychological therapies

Depression and other psychiatric diagnoses are common in SCD, with contributing factors including illness chronicity, the unpredictable nature of crises and various medical complications (Molock & Belgrave, 1994). With thalassaemia now a chronic condition, focus has shifted to the recognition and management of psychosocial problems resulting from the condition and its intensive and demanding treatment regimes (Var-daki et al., 2004). Thus, clinical psychology can impact service user experiences and effective care delivery.

To highlight the potential of having a CP on the team, short-term psychological therapies and self-hypnosis were trialled with five individuals by a Consultant CP (DM) qualified to practice hypnotherapy. This was found to be most effective during in-patient admissions or when run alongside haemoglobinopathy outpatient clinics. Research at other centres has found psychological interventions, such as cognitive behavioural therapy, to be both clinically and cost effective (Thomas et al., 2001). Relaxation and self-hypnosis in SCD helps decrease stress and anxiety, which are known to precipitate and exacerbate crises (Dinges et al., 1997). Service user and healthcare professional feedback was very positive and it was clear that psychological approaches would be accepted as an adjunctive therapy for managing and preventing painful crises.

Neuropsychology in SCl)

Neuropsychological deficits resulting from clinical or silent strokes in SCD have been documented in the paediatric population (Berkelhammer et al., 2007). Impairments in executive function and attention are common indicators of cerebrovasculopathy in SCD (Kral et al., 2003). Such impairments can affect an individual's ability to access and engage with services and complex care regimes across the lifespan. In the UK, it is recommended that all children with the most severe form of SCD, haemoglobin SS (HbSS) are screened annually using transcranial doppler (TCD) scanning to assess the risk of stroke. Although MRI scans can detect silent infarcts, it is impracticable and expensive to scan every child regularly. Thus, it is recommended that regular developmental assessments and neuropsychological screening be conducted.

Children with abnormal TCD results have decreased verbal intelligence and executive function compared to children with normal or conditional results (Kral et al., 2003). Neuropsychological assessment of large numbers of children, however, is time-consuming and costly. Initially, we were uncertain of how many high-risk children (with abnormal TCD results) would be referred for assessment in a year-long period. This project enabled us to examine the feasibility of testing those with abnormal scans over this period with the Wechsler Intelligence Scale for Children (WISGIV). By completing these tests, we were able to demonstrate the feasibility of administering full intellectual assessments to the small number of high-risk children (n = 2). The assessments also offered a baseline measure, so that pre-morbid measures were available should these children suffer strokes in the future.

The literature on adult cognitive deficits in SCD is sparse but suggests that neuropsychological impairment in some adults is likely (Vichinsky et al., 2010). We assessed two adults with SCD who presented with cognitive difficulties impacting on their daily life and care regime. Assessments were tailored to each individual's presenting complaints. We found the intellectual function of one adult to be within the 'Extremely Low' range on the Wechsler Adult Intelligence Scale (WAIS-IV; Full Scale .IQ = 57). Pre-morbidly, this patient was estimated to have functioned in the

average to low-average range. Further assessments completed by colleagues in the adult neuropsychology service found low results in numerous domains including executive function and short-term memory; however, interpretation was complicated by variable engagement across the assessment. The results were fed back to the care team who interpreted them alongside an MRI scan which found deep white matter lacunar infarcts. Lacunar infarcts have been found in SCD (Hoppe, 2005) and are associated with neuropsychological impairment, which can worsen over time (Samuelsson et al., 1996).

The second adult did not show MRI abnormalities, but performed at the 5th percentile overall on the Doors and People Test and below the 5th percentile on the Rivermead Behavioural Memory Test. The Doors and People test revealed poor immediate recall whilst forgetting scores were only just below average. This may suggest attentional problems or executive dysfunction which has been implicated in SCD (Kral et al., 2003). Despite these neu-ropsychological impairments, no MRI abnormalities were found when investigated. This finding is supported by Vichin-sky et al. (2010), who suggest that neurologically intact SCD patients can present with neuropsychological deficits. Our assessments provided the team with important information which enhanced care delivery by confirming the impairments and identifying ways in which to improve engagement and understanding. The results suggested the need for a wider investigation of neuropsychological impairments in the adult SCD population, which would inform local needs and enhance the evidence base.

Focus group findings

The focus group provided service user perspectives to our case of need. Four individuals with SCD attended the discussion, which lasted approximately one hour. Informed consent was obtained from participants who agreed for anonymous quotes to be used in project outputs.

The focus group discussion was facilitated by the AP, using a semi-structured interview schedule (developed by the team). It was audio recorded and transcribed verbatim. The transcript was thematically analysed by RdN and **JL,** who independently coded the data and came to a consensus on the coding and thematic structure. Three main themes emerged from the focus group which were: existential concerns, stigma, and the need for alternative treatments. Here, we describe the latter theme, which informs practice and was a common theme throughout the discussion. In these instances, service users explicitly referred to aspects of their care that could be improved by the inclusion of a clinical psychology service. The theme is divided into the following sub-themes...

Dealing with existential issues

Service users spoke about life and mortality, with thoughts about death becoming so commonplace that there was some ambivalence about how to deal with this.

> R3: When I was a kid... I used to be the most maddest... used to go on all sorts of trips and be in all types of cold water 'n' stuff - never ever suffered from it. And then got older and older and then now it's just getting worse and worse. And I've started thinking to myself now, how old am I going to be when **I** die?
>
> **RI:** Ahl I do that. I think about that all the time.
>
> R4: I'm like: 'Well I might never live past sixty anyway.' [laughs]

While it appears that they use humour to cope with this uncomfortable thought, service users felt the need to have someone they could talk about their fears to. This is exemplified here:

> R2: So if there was a service around that where, you know, sickle cell people can actually go and talk to someone about their fears about that [death and dying]. That will help them as well.

Medication

While all service users had some reservations about medication use to control the condition, and the way in which they were prescribed, all understood and appreciated the need for taking some, particularly during severe crises. Some tried to avoid taking drugs as long as they could help it, but at the point of crisis, they did not want other forms of intervention:

> **RI:** No, I mean I wouldn't try anything for my crises [because] crises that I'd have to go into hospital for I'd definitely, you know, because I want it [drugs] [snaps finger] in an instance. I wouldn't have the patience to go through breathing exercises [R2 & R3 laugh] when you're in pain. I just want something there and now [laughs].

However, when the pain had reduced, they were less keen to continue taking medications, as they often had distressing side-effects which they felt were not acknowledged by clinicians.

> R3: You're drugging me and not asking me how these drugs are making me feel. I don't know... No, not once have they ever asked me 'How does these tablets make you feel? How does Tramadol make you feel? How does Cycliyzine make you feel? Paracetemol? Diclofenac?'. Never once have they asked me how the drugs make you feel. Cause it's all right them prescribing you drugs and pumping you full of drugs. [But] then you're mind ain't right. And you're body ain't right...

The potential of clinical psychology R3 spoke positively about an imagery technique she had been taught as part of our project:

> R3: Well she [CP] came, this is a while ago now when I was in hospital, and she sat beside my bed... And she did this - close your eyes breathe, look for this box... whole thing... That kinda helped, cause the pain was still there but it was kind of far away. Sometimes your pain just feels like it's there but it's distant. That helped.

A final excerpt summarises the potential of a clinical psychology service for dealing with the complex difficulties that this population face:

> R1: So the psychology service addresses the fact that we feel like frauds, it's an invisible disease, nobody understands and we're not believed. We're not believed whenever something is wrong with us, and we're constantly having to make life-changing decisions at such a young age, and we always have to worry about when we're going to die or when we are going to get stroke...

By voicing these experiences, we were able to directly represent service users' views and opinions to commissioners and treating teams. The focus group findings were therefore a key piece of evidence in our bid for funding.

The toolkit

Throughout the project we developed a toolkit of materials to enable a newly appointed CP to start work promptly. The toolkit included:

- a guide to local haemoglobinopathy services;
- standards and guidelines;
- clinical psychology leaflet for service users;
- referral forms;
- psychological screening tools;
- a transition plan (from paediatric to adult services); and
- staff training materials (e.g. painful procedures).

Conclusions

Our project concluded with the submission of a case of need to the local NHS Specialist Commissioning Group. A meeting was then held with the commissioning group's Strategy and Planning Team who, following our work, confirmed funding for a CP to work within the team. This project was unique in that it enabled substantial groundwork to be completed prior to the implementation of the service. Discussion with others working in the speciality and

previous work by Thomas et al. (2009) has highlighted the value of conducting this type of groundwork prior to setting up and delivering a service. Therefore, by employing an AP for a time-limited period, we were able to set up the service in a cost-effective manner.

In haemoglobinopathy services clinical psychology equips people with the skills required to cope effectively with their condition, with the aim of reducing hospital admissions, increasing adherence, and improving quality of life. Our service evaluation identified the key issues and needs of this population, and allowed us to establish links and introduce clinical psychology to the team and service users. This process facilitated a straightforward bid for funding, which proved a success.

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